An extra hill in a poly-hill sign in a patient with facioscapulohumeral dystrophy

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An 18-year-old boy presented in the outpatient clinic with difficulty in raising hands above shoulders and inability to sip drinks through a straw. Examination revealed marked weakness of the muscles around shoulder girdle, particularly trapezius, serratus anterior, biceps and triceps muscles. Mild weakness was noted in facial, proximal lower limb and anterior tibial muscles. The scapulae were winged and placed wide apart. His serum creatine kinase level was marginally raised at 510IU/L. Electromyography showed myopathic pattern. A clinical diagnosis of facioscapulohumeral dystrophy (FSHD) was made. As described earlier, poly-hill sign was elicited by asking patient to raise his arms with elbows flexed to nearly 90° and shoulders abducted to his maximum ability (around 70°).[1] The sign [Figure 1] which showed six “hills” on either side consisted of (1) enlarged infraspinatus muscle overlying the winged inferior angle of scapula, (2) upwardly projected superior angle of scapula tenting the wasted trapezius muscle, (3) prominence of laterally projected acromioclavicular joint due to wasting of trapezius muscle attached to it medially and supero-medial part of deltoid muscle attached to it laterally, (4) prominence of inferolateral part of the deltoid muscle due to wasting of its own supero-medial part on one side and biceps brachii muscle on the other side, (5) unusual bulge due to some preserved bulk in the middle of the wasted biceps brachii muscle and (6) prominent and well preserved extensor digitorum communis / brachioradialis muscles. The poly-hill sign is based on the fact that various muscular dystrophies have selective weakness, wasting or enlargement of either a group of muscles or a part of a muscle.[2,3] Patients with FSHD generally have markedly wasted biceps brachii muscle resulting in the description of arm’s appearance as “Popeye’s arm”. [4] Classical “poly-hill sign” therefore has only five hills [Figure 2]; bulge in the middle of the biceps brachii muscle as seen in this patient, is an unusual feature.

![Figure 1: Photograph of a patient with facioscapulohumeral dystrophy demonstrating a poly-hill sign with an extra hill over the biceps muscle](image1)

![Figure 2: Photograph of a patient with facioscapulohumeral dystrophy demonstrating a classical poly-hill sign](image2)
The poly-hill sign is most useful in the diagnosis of FSHD in patients with minimal or insignificant facial muscle involvement

References


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